CASE REPORT

Jugulotympanic Paraganglioma, Mimicking Chronic Suppurative Otitis Media

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ABSTRACT

Paraganglioma or glomus tumour are named according to their origin. Jugulotympanic paragangliomas (JTP) originates in the middle ear. There are several diagnostic pitfalls of this tumour. We now report a rare case of JTP in a 47 years old female. Pre-operative diagnosis of this case was chronic suppurative otitis media, Per-operatively it revealed an irregular somewhat polypoid mass. On histologic examination the mass composed of nests of round to oval cells surrounded by delicate vascular septae and the diagnosis was JTP. Because JTPs are rare tumour and have variable clinical presentation & different histologic findings they are easy to misdiagnose. However this case report may help to generate awareness and to avoid misinterpretation of JTPs.

Keywords: Paraganglioma, Jugulotympanic, Middle ear

Introduction

The term paraganglion was first used by histologist Kohn in the early part of this century to describe the carotid body[1]. Paragangliomas in the head and neck are relatively rare tumours arise at four sites: the carotid body, the middle ear, the jugular bulb and the vagus nerve. Among them, jugulotympanic paragangliomas (JTPs) arise from anatomically dispersed paraganglia near the base of the skull and middle ear. Cuild first described vascularized tissue in the dome of the jugular bulb and on the promontory of the middle ear and named as "glomic tissue" in 1941[2]. The incidence of jugulotympanic paragangliomas is approximately 1:1,300,000[3]. Under microscopic examination, JTPs tend to be more vascular, and cell nests are less uniform and frequently smaller compared with other paragangliomas. Dense sclerotic matrix is another characteristic of JTPs. Vascular tumor, middle ear adenoma and even a reactive condition such as granulation tissue in chronic otomastoiditis are diagnostic pitfalls of this tumor. Therefore during interpretation of a tumor presenting as a middle ear or temporal bone mass, this pitfall should be kept in mind. But pulsatile tinnitus is a very important clinical findings to differentiate glomus tumour from others.

Case Report

A 47 year old female was admitted with the history of earache, right sided ear obstruction and hearing disturbance for the last eight years. She also complained about discharge of pus and bleeding from right ear for the last 6 months. On physical examination, the right external auditory canal was narrowed and was clogged by crust with an invisible tympanic membrane. Audiometry reveals conductive type of deafness. X-ray findings not available. Other routine examination was within normal range. Provisional diagnosis was chronic suppurative otitis media. Patient treated conservatively without any improvement. Finally mastoidectomy was planned. After a retroauricular incision and open cavity tympanomastoidectomy, an irregular somewhat polypoid mass with a bleeding tendency was found, that filled the external auditory canal, middle ear cavity and mastoid process. Whereas the tympanic membrane was not found, and auditory ossicles were disrupted, the sigmoid sinus, dura matter and facial canal were intact. Most of the mass was removed, but some remained because of massive bleeding. On histologic examination, highly vascular tissue looking like granulation tissue was seen in a low

power microscope field. In high power field it reveal that the tumour is composed of nests of round to oval cells that are surrounded by delicate vascular septae resembling zellballen appearance. The tumour cells contain granular eosiophilic cystoplasm and uniform round to ovoid nuclei (Fig 1).

Although considerable squeezing artifacts were present, this case was diagnosed as a jugulotympanic paraganglioma histologically.

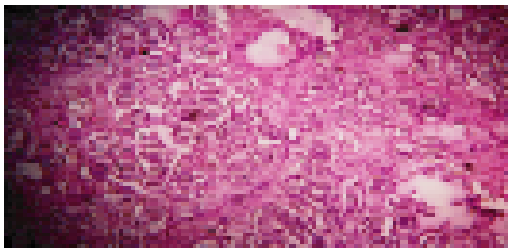

Figure 1: *Histological section showing tumour composed of nests of round to oval cells that are surrounded by delicate vascular septae resembling zellballen appearance.(H&E×100)*

Discussion

JTPs can be confused with vascular tumor, middle ear adenoma and even with a chronic otomastoiditis which occurred in this case. The two entities-glomus tumors and JTPs that are called "glomus" tumors, are definitely different diseases. JTPs are tumors arising from paraganglia whereas glomus tumors arise from a modified smooth muscle cell located in the walls of specialized arteriovenous anastomoses involved in temperature regulation.

The major differential diagnosis of JTPs includes ruling out a vascular tumor such as an epithelioid hemangioendothelioma (EHE) because of the high vascularity of JTPs. EHE can be differentiated from JTPs by the findings in tumor cells having moderate amount of pale eosinophilic cytoplasm and immunore-activity for vascular markers such as CD3l, CD34 and factor VIII[4]. We also thought this case was a vascular tumor, because there were numerous small-sized vessels and the small cells between the vessels. There are some reported cases of jugulotympanic paraganglioma which mimics vascular tumor[5]. However, through careful microscopic examination, the mass was shown to be a neuroendocrine tumor. Another important alternative in the differential diagnosis is middle ear adenoma (MEA), which is thought to arise from pluripotent cells in the middle ear mucosa and may have mixed patterns of differentiation, ranging from glandular to neuroendocrine[6]. Because these tumors may have a neuroendocrine component, MEAs can be confused with paragangliomas. However, less vascularity and mixed patterns of differentiation of MEAs may be helpful to differentiate it from paragangliomas. JTPs usually develop in adults in the 5th to 6th decade of life & unlike carotid paraganglioma there is a definite female predominance in the incidence of jugulotympanic paniganglioma with a female to male ratio of 4:13. In this case, the patient was a female aged 47 yrs. Recently, published clinical review of about 9 Korean cases of jugular paraganglioma[7] (Table 1) is given below along with our case.

Table I: Clinical presentation of 9 Korean cases of jugular paraganglioma[7], Chung g et al and our case.

Case No.					Initial symptom
1.	F	32	Left	1.5×1	Pulsatile tinnitus, headache
2.	M	35	Right	3x3	Facial palsy
3.	M	26	Left	3x2	Pulsitile tinnitus, otalgia
4.	M	48	Left	2.2 x 2	Hoarseness
5.	M	45	Left	4.5 x 3.5	Hoarseness, chronic cough
6.	F	50	Right	2.2 x 25	Sensorineural hearing disturbance
7.	F	60	Right	4.5 x 2.2	Hoarseness
8.	F	29	Right	4.5 x 9	Hoarseness, facial palsy
9. Ji-YounSung et al	M	18	Right	2.7x 2.5	Sensorineurla hearing loss, pulasitile tinnitus
10. Our case	F	47	Right	1.5 x 1	Pulsatiletinnitus, pus discharge, bleeding.

These review included 4 men and 5 women who underwent surgery between 1986 and 2005. Mean age at the time of diagnosis was 40.8 years (range, 26 to 60 years).

Conclusion

JTP is a rare tumor and can be missed or misinterpreted due to its different histology. Keeping in mind about this entity and its characteristics, it would be helpful in leading to a correct diagnosis when we interpret middle ear or temporal bone masses.

References

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